Don’t Be Blind to the Diagnosis: An Intriguing Case of Monocular Blindness

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Don't Be Blind to the Diagnosis: An Intriguing Case of Monocular Blindness

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Introduction

Vasculitis is a broad term for conditions involving the inflammation of blood vessels. They are typically characterized by the size of vessels involved. For instance, giant cell arteritis (GCA) is a large vessel vasculitis commonly seen in the temporal artery and the most feared complication is acute vision loss. Another vasculitis, granulomatosis with polyangiitis (GPA) is a small-to-medium sized vasculitis most commonly associated with c-ANCA positivity and is characterized by necrotizing granulomas. Differentiating between various types of vasculitides can prove to be diagnostically challenging.

Case Description

A 67-year-old female with history of sero-negative rheumatoid arthritis and newly developed lower extremity ulcerations (Figure 1) presented to the hospital with acute painless vision loss of the right eye. She was found to have an elevated sedimentation rate of 122. Ophthalmology diagnosed the patient with central retinal artery occlusion (CRAO) thought to be secondary to either rheumatoid vasculitis or GCA.

The patient was placed on pulse steroids and underwent temporal artery biopsies. Her stay was complicated by the development of left hand and right foot drop, proteinuria, and acute kidney injury. Her hand and foot drop were evaluated with electromyography and was consistent with mononeuritis multiplex. Biopsy of her lower extremity lesions only revealed panniculitis. Eventually, temporal artery biopsy came back positive for inflammatory changes in the smaller branches of the temporal artery, which could be consistent with GCA in the correct clinical picture (Figures 2 and 3). However, due to the patient's multitude of symptoms, further work up was pursued. Laboratory evaluation revealed high titer PR3 antibodies and she underwent renal biopsy which revealed pauci-immune glomerulonephritis, most compatible with PR3 ANCA vasculitis/GPA. She underwent treatment with cyclophosphamide and rituximab and achieved remission; however, did not regain sight in her right eye.

Discussion

In this patient's case, her presentation of temporal arteritis and inflammation seen on biopsy was not true giant cell arteritis but part of a larger diagnosis of PR3 ANCA vasculitis. Failure to pursue further work up after finalization of the temporal artery biopsy would have led to a missed diagnosis and inappropriate treatment. CRAO is a previously reported, however quite rare, manifestation of GPA. Treatment of high dose steroids only would have been detrimental and possibly even fatal in this patient. It is therefore important to only stop investigating once the diagnosis appropriately fits the overall clinical picture.

Figure 1. Right lower extremity skin lesion.

Figure 2. Normal section of temporal artery.

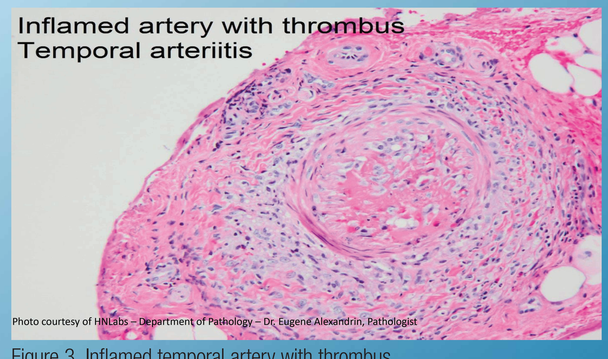

Figure 3. Inflamed temporal artery with thrombus.

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